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Case Report

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Pheochromocytoma in pregnancy: A case report

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Introduction

Pheochromocytoma is a rare tumor derived from chromaffin tissue which secretes large amounts of catecholamines, giving rise to the characteristic signs and symptoms of this condition. The tumor poses a major challenge to the anesthesiologist due to the unstable and unpredictable clinical condition of the patient, both during anesthesia and surgery. 85% of pheochromocytomas are located within the adrenal medulla, and 98% are within the abdomen. Extra adrenal peochromocytomas or paraganglionomas may occur anywhere in the body where there sympathetic nervous tissue, such as the abdominal or thoracic sympathetic chain, bladder wall heart, mediastinum, carotid and glomus jugulare bodies. The overall incidence of the tumor is between 1.55 and 2.1 per million per year.[1,2] The association with pregnancy and pheochromocytoma is extremely uncommon, with an estimated incidence of 0.2 per 10,000 pregnancies.[3] Being a high risk condition for the both the mother and the fetus, successful outcome depends on early diagnosis, adequate therapy, and decision when to undertake surgery during pregnancy.

The definitive management of this condition is surgery and, with developments in laparoscopic surgery, this is undoubtedly the ideal approach for tumor resection.[4] The benefits of laparoscopic technique over conventional open surgery are smaller incision, less post operative pain, shorter hospital stay and low perioperative morbidity. We present the case of a patient with 25 weeks of gestation with a diagnosis of pheochromocytoma and treated with laparoscopic surgical resolution.

Case report

Our patient, a 30 year old third gravida at 23 weeks gestation, with history of two previous cesarean sections, was diagnosed with pheochromocytoma of the right adrenal medulla two years ago. She had clinical features of palpitations. tremors and profuse sweating with sinus tachycardia and elevated blood pressure values. Laboratory investigations showed hyperglycemia and elevated 24 hour urinary metanephrines. Ultrasound abdomen showeda right adrenal tumor. After an interdisciplinary medical board composed of I.T.U, surgery, gynecology, urology, endocrinology and anesthesiology services; it was decided to undertake laparoscopic removal of tumor at 25 weeks gestation. Two weeks prior to her surgery, the patient was treated with 14 units actrapid insulin subcutaneosly twice a day , tablet doxazosin 6mg orally twice dailyand her blood pressures touched normal after two days. Treatment was continued and, three days before the surgery, tablet carvedilol 12.5mg twice daily was added.

Her preoperative examination revealed pulse rate of 78/min, blood pressure 121/78mmHg, oral temperature of 36.5 °C, capillary glucose 176 mg/mL. Fetal heart rate was 140/min. Bispectral index was 96%. A bolus dose of 2 g of Inj magnesium sulfate was administered intravenously over a period of 20 minutes and an epidural catheter was placed at L1-L2 interspace without complications. Patient was premedicated with 3.5 mg midazolam intravenously. After a negative Allen test, a left radial arterial line was established with a 20 gauge cannula for invasive blood pressure monitoring.

After 5 min preoxygenation, an infusion of sodium nitroprusside (SNP) at a rate of 0.5mcg-kg/min was initiated to control the hypertensive response to intubtion and anesthesia induced with an infusion of remifentanyl at a rate of 0,1 mcg-Kg/min, Inj lidocaine 60 mg bolus followed by 90 mg of and propofol. Tracheal intubation using videolaryngoscopy with a 7 mm ID cuffed endotracheal tube 30 seconds following 36 mg rocuronium intravenously was achieved. The right internal right jugular vein was cannulated without complications to monitor cvp.

Anesthesia was maintained sevoflurane 1.3-1.8 vol% and remifentanyl 0.1-0.5 mcg-Kg/min. The SNP infusion at 0.5-5mcg / kg/minute was continued to maintain blood pressure values within normal limits. Manipulation of the tumor resulted in supraventricular tachycardia episodes (140-150 bpm) when two doses of 6 mg adenosine normalized the heart rate. Pressure controlled ventilation with equal flows of oxygen and air was done to maintain Pet CO_2 value between 30-35 mmHg. Just before ligation of the tumor pedicle, SNP infusion was discontinued and volumetric expansion carried out with 600 ml lactated Ringer's. Patient hemodynamics remained stable there was no need for vasopressors.

Prior to skin closure, epidural top up of 10 ml 0.1% bupivacaine 0.1% with 25 microgram fentanyl was made through the epidural catheter for post operative analgesia. When patient's bispectral index was 86, 120 mg sugammadex was administered for reversal of neuromuscular blockade and patient extubated. Doppler examination revealed fetal vitality and the patient was subsequent transferred to the intensive care unit.

Discussion

Pheochromocytoma with pregnancy is an extremely rare entity and the decision to undertake surgical removal of the tumor has to be made based on several factors like the gestational age and normal clinical and biochemical parameters. It is generally agreed that the end of the second quarter, (weeks 24 and 25) is the ideal time for

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extirpation surgery.[5] The appropriate preoperative management of pheochromocytoma prior to surgery with alpha and beta blocker drugs and also the optimization of blood sugars, plays an indispensable role for the successful outcome of these patients.[6] In this regard the efforts are focused on the following goals:

a. Blood pressure control with alpha blockers.[7] (phenoxybenzamine, doxazosin) for a fortnight before surgery.

b. Control of heart rate and arrhythmias. This should be done only after achieving blood pressure control.[8] Calcium channel blockers with or without beta blockers can control cardiac rhythm abnormalities successfully.

c. Restoration of contracted circulating volume under cvp monitoring is recommended.

Is globally accepted that the laparoscopic technique is the best surgical approach for resection of pheochromocytoma, as it is safe and effective, minimally invasive and is associated with a faster recovery and shorter hospital stay.[9,10] In our case the laparoscopic transperitoneal lateral approach was decided, taking into account the size of the pregnant uterus, keeping lower intra abdominal pressures of 12 mmHg during pneumoperitoneum and also lower CO2 gas flows of 10 L/min to minimize the surge in catecholamines caused by hypercapnia.[11]

The pregnant patient scheduled for resection of pheochromocytoma presents additional challenges for anesthetic management, like fetal acidosis during carbon dioxide insufflation into the maternal peritoneal during laparoscopic surgery.[12] The transfer of CO2 through the placenta has not been associated with long-term adverse effects in the fetus, newborn or child.[13] An additional concern is the threat of premature delivery or risk of abortion due to the surgical manipulation.[14] In our case, the use of magnesium sulfate[15] was made to reduce that risk.

Anesthetic induction and maintenance must aim at minimizing hypertensive crisis. This calls for strict vigilance over the endpoints of greatest hemodynamic stress. In this regard is imperative to maintain control of blood pressure values during laryngoscopy, intubation and surgical manipulation of tumor.[16,17] Administration of direct vasodilators such as sodium nitroprusside (SNP), nitroglycerin and urapidil are recommended. The appearance of supraventricular arrhythmias usually occurs during tumor manipulation.[18] We opted for the administration of intravenous esmolol at doses of 6.0 mg.

Another aspect of fundamental importance is the maintenance of adequate depth of anesthesia. Light planes of anesthesia coupled with surgical stimulation will result in a sympathetic response in the patient and trigger a hypertensive crisis. We avoided this by monitoring the bispectral index[19] and ensuring balanced general anesthesia coupled with neuroaxial block to prevent this with success.[20] Hypotension is frequently seen after clamping the venous drainage of the tumor. Vascular expansion with crystalloid solutions, prior to the ligation of the vein and administration of small doses of phenylephrine or continuous infusion of norepinephrine may be required to prevent this.[21]

Conclusion

We report a case of a patient with 25 weeks of gestation and a diagnosis of pheochromocytoma who was successfully managed with laparoscopic adrenalectomy. Good team work, optimal preoperative preparation and judicious anesthetic management under evidence based monitored care had a successful outcome without compromising the featl well being. The patient was discharged on the seventh post operative day and went on to term pregnancy and normal delivery of a healthy baby.

Conflicts of interest

There is no conflict of interest.

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